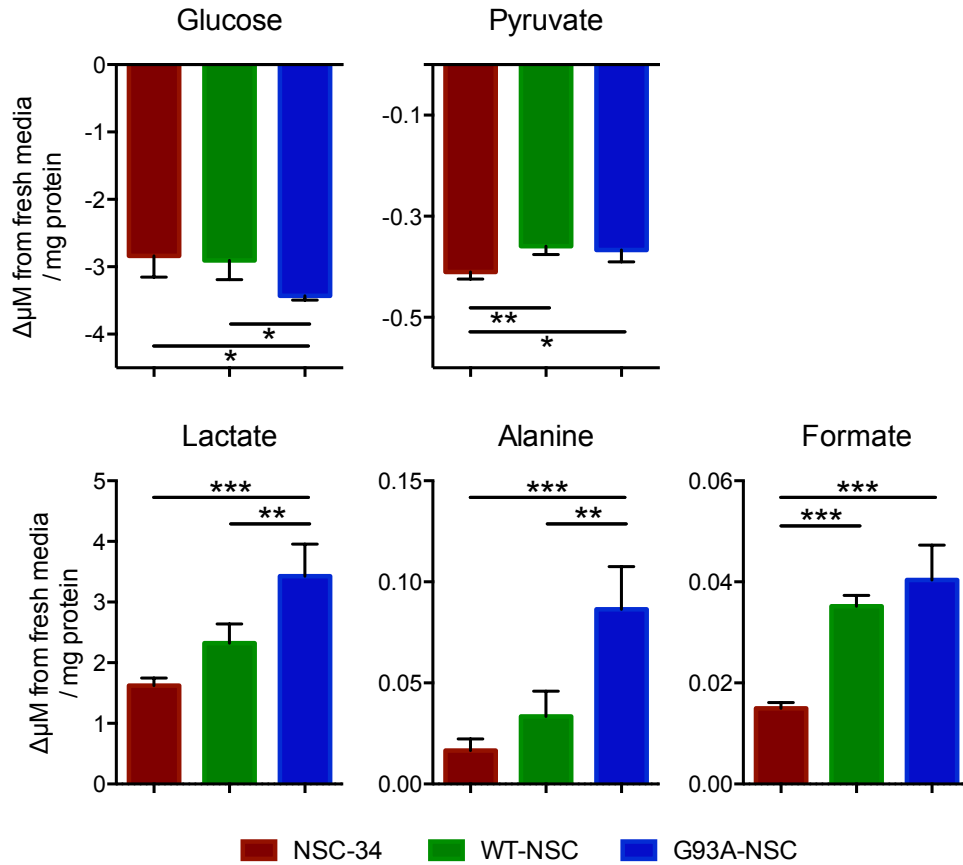
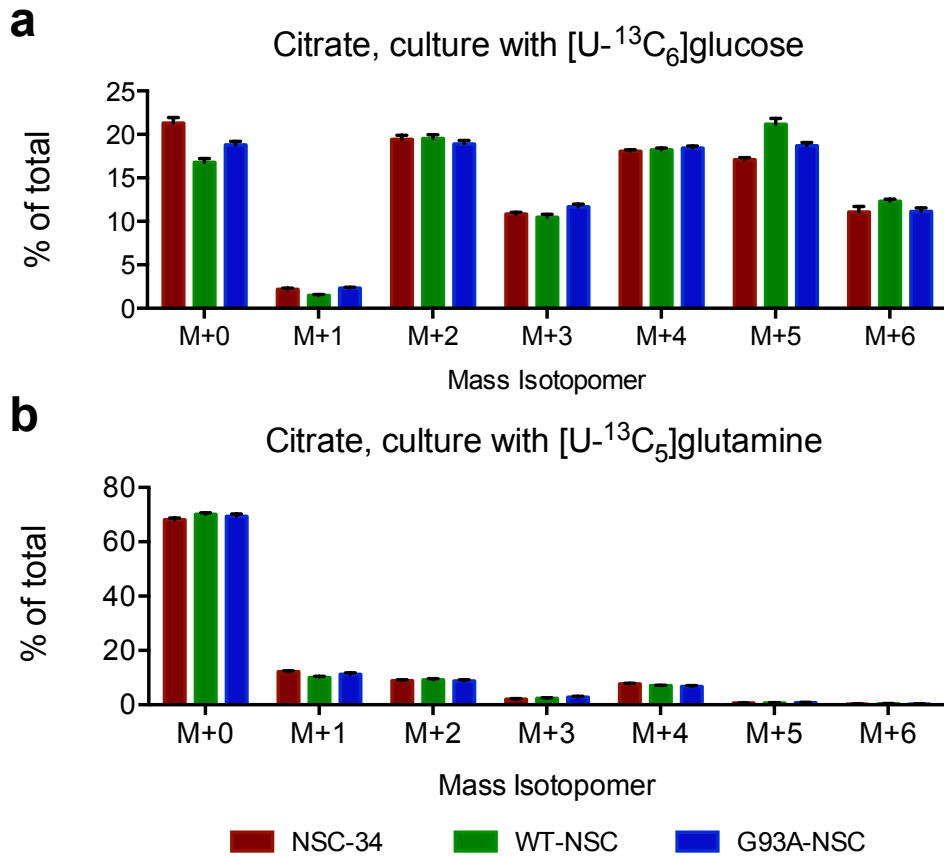


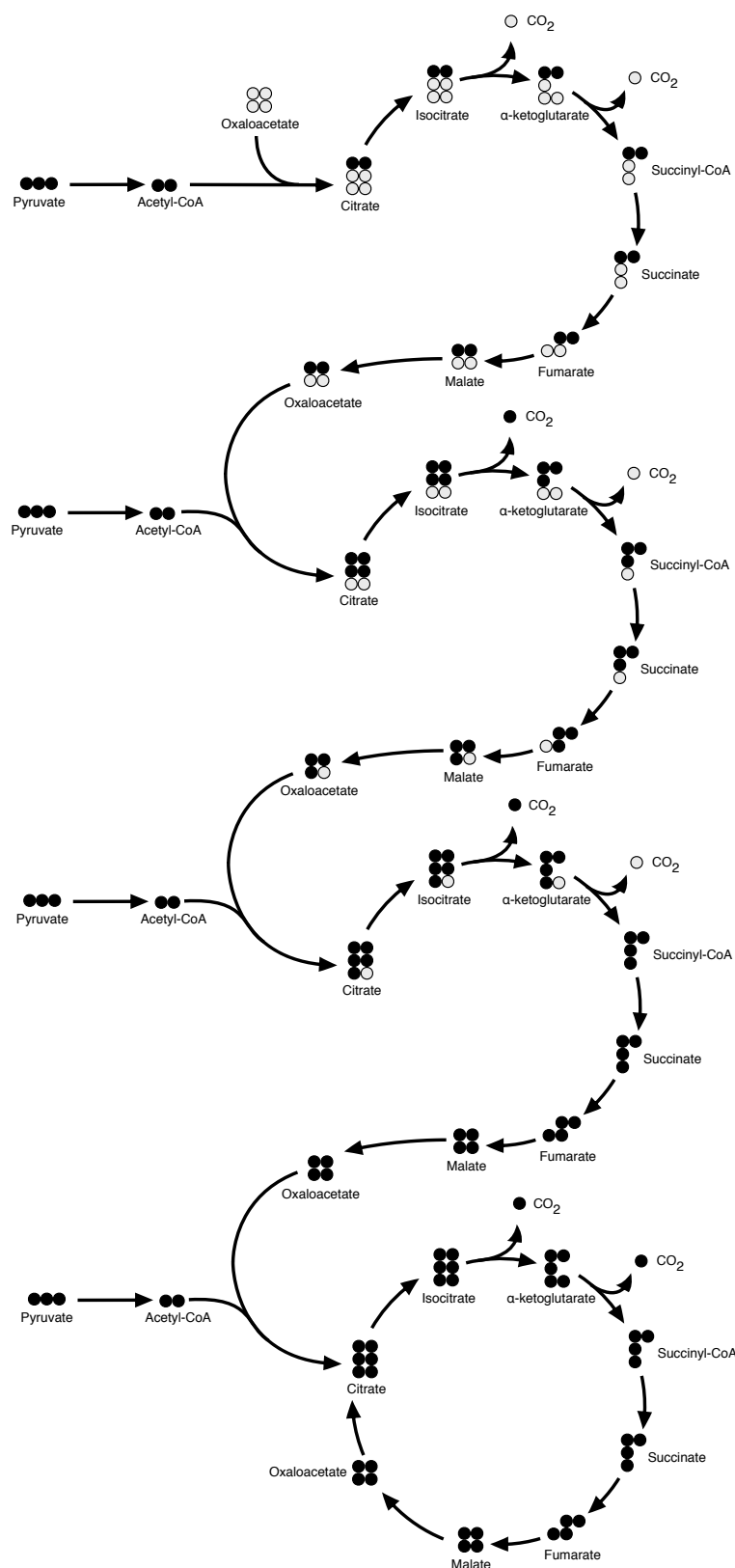
SUPPLEMENTARY FIGURES



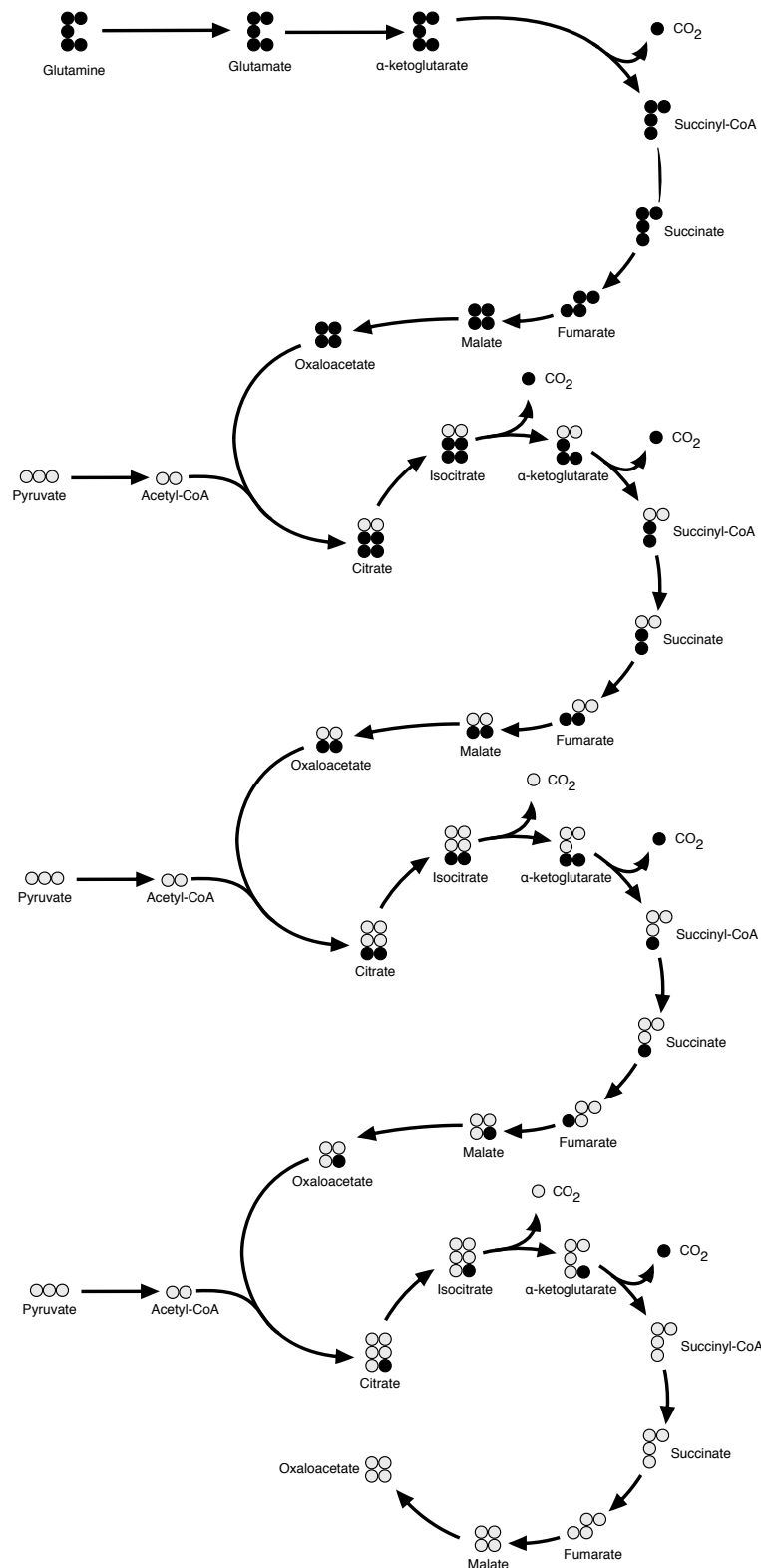
Supplementary Figure 1. Extracellular metabolome characterization reveals regulation of glycolysis with expression of wt- and G93ASOD1 under culture with 5% FBS. The NSC-34, WT-NSC and G93A-NSC cell lines were cultured for 22 h with 5% FBS, the media was collected and metabolites were determined by ^1H NMR spectroscopy. All values are mean \pm s.e.m. (n=4, * p<0.05, ** p<0.01, *** p<0.001 after one-way ANOVA with False-Discovery Rate (FDR) correction for multiple testing and Tukey's post-hoc test).



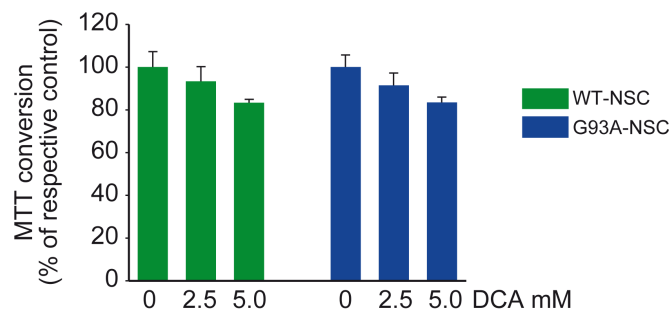
Supplementary Figure 2. Effect of wt- and G93ASOD1 expression on the mass isotopomer distribution of citrate after culture with labeled glucose or glutamine. The NSC-34, WT-NSC and G93A-NSC cell lines were cultured for 22 h without serum and with [U-¹³C₆]glucose or [U-¹³C₅]glutamine. Histograms show the mass isotopomer distribution of citrate after culture with (a) labeled glucose or (b) labeled glutamine. All values are mean \pm s.e.m. (n=5).



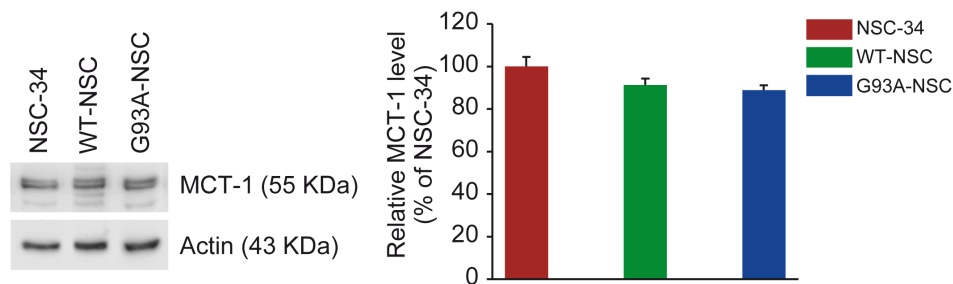
Supplementary Figure 3. Labeling scheme from U-¹³C pyruvate entry into the TCA cycle after culture with [U-¹³C₆]glucose. Potential mass isotopomers for TCA cycle intermediates after entry of [U-¹³C₃]pyruvate from [U-¹³C₆]glucose in repeated cycling through the TCA cycle.



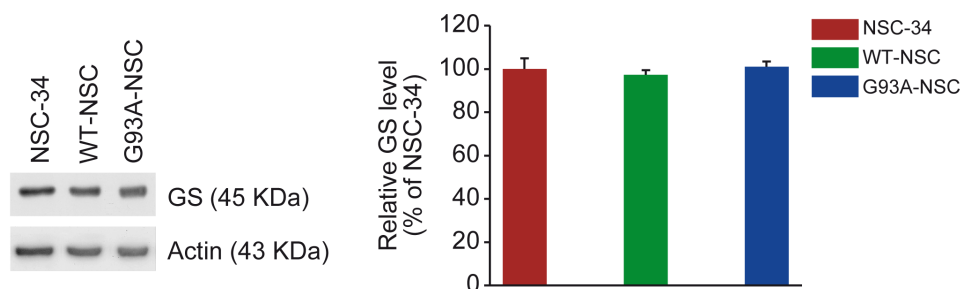
Supplementary Figure 4. Labeling scheme of ^{13}C incorporation into the TCA cycle after culture with $[\text{U-}^{13}\text{C}_5]\text{glutamine}$. Potential mass isotopomers for TCA cycle intermediates after labeling with $[\text{U-}^{13}\text{C}_5]\text{glutamine}$ and subsequent dilution of ^{13}C with the entry of ^{12}C pyruvate in repeated cycles.



Supplementary Figure 5. Viability of the WT-NSC and G93A-NSC cell lines after treatment with sodium dichloroacetate. The WT-NSC and the G93A-NSC cell lines were cultured without serum for 46 h with/without sodium dichloroacetate (DCA) 2.5mM and 5mM. Viability of the cell lines was determined with the MTT assay. Values (mean \pm s.e.m.) are percentages of the MTT conversion after culture without DCA (100%) (n = 3).



Supplementary Figure 6. Expression level of the monocarboxylate transporter 1 protein of the NSC-34, WT-NSC and G93A-NSC cell lines. The NSC-34, WT-NSC and G93A-NSC cell lines were cultured without serum for 22 h and the levels of monocarboxylate transporter 1 (MCT1) protein were determined by Western blot and normalized to actin. The value of the NSC-34 cell line was taken as 100%. Means \pm s.e.m. (n=6) and a representative Western blot are shown.



Supplementary Figure 7. Expression level of glutamine synthetase protein of the NSC-34, WT-NSC and G93A-NSC cell lines. The NSC-34, WT-NSC and G93A-NSC cell lines were cultured without serum for 22 h and the levels of glutamine synthetase (GS) protein were determined by Western blot and normalized to actin. The value of the NSC-34 cell line was taken as 100%. Values are shown as mean \pm s.e.m. (n=3) and a representative Western blot is presented.